SANTA CRUZ BIOTECHNOLOGY, INC.

AGL (B-11): sc-518176

SANTA CRUZ BIOTECHNOLOGY

The Power to Question

BACKGROUND

AGL (amylo-1, 6-glucosidase, 4-α-glucanotransferase), also known as GDE (glycogen debranching enzyme), is a 1,532 amino acid protein that exists as three alternatively spliced isoforms which are expressed in kidney, liver, heart and muscle in an isoform-specific manner. Exhibiting multifunctional enzyme capabilities, AGL contains two catalytic active sites, one of which acts as an 4-α-glucotransferase and the other of which acts as an amylo-1,6-glucosidase during glycogen degradation. Defects in the gene encoding AGL are the cause of glycogen storage disease type 3 (GSD3), also known as Forbes disease. GSD3 is a metabolic disorder that is characterized by the presence of abnormal glycogen due to a lack of AGL activity. Symptoms of GSD3 generally include hypoglycemia, variable myopathy, hepatomegaly and short stature.

REFERENCES

1. Ding, J.H., et al. 1990. Immunoblot analyses of glycogen debranching enzyme in different subtypes of glycogen storage disease type III. J. Pediatr. 116: 95-100.
2. Yang, B.Z., et al. 1992. Molecular cloning and nucleotide sequence of cDNA encoding human muscle glycogen debranching enzyme. J. Biol. Chem. 267: 9294-9299.
3. Shen, J., et al. 1996. Mutations in exon 3 of the glycogen debranching enzyme gene are associated with glycogen storage disease type III that is differentially expressed in liver and muscle. J. Clin. Invest. 98: 352-357.
4. Orho, M., et al. 1998. Mutations in the liver glycogen synthase gene in children with hypoglycemia due to glycogen storage disease type 0. J. Clin. Invest. 102: 507-515.
5. Horinishi, A., et al. 2002. Mutational and haplotype analysis of AGL in patients with glycogen storage disease type III. J. Hum. Genet. 47: 55-59.

CHROMOSOMAL LOCATION

Genetic locus: AGL (human) mapping to 1p21.2; Agl (mouse) mapping to 3 G1.

SOURCE

AGL (B-11) is a mouse monoclonal antibody raised against amino acids 1233-1532 mapping at the C-terminus of AGL of human origin.

PRODUCT

Each vial contains 200 µg IgG_1 kappa light chain in 1.0 ml of PBS with < 0.1% sodium azide and 0.1% gelatin.

AGL (B-11) is available conjugated to agarose (sc-518176 AC), 500 µg/0.25 ml agarose in 1 ml, for IP; to HRP (sc-518176 HRP), 200 µg/ml, for WB, IHC(P) and ELISA; to either phycoerythrin (sc-518176 PE), fluorescein (sc-518176 FITC), Alexa Fluor® 488 (sc-518176 AF488), Alexa Fluor® 546 (sc-518176 AF546), Alexa Fluor® 594 (sc-518176 AF594) or Alexa Fluor® 647 (sc-518176 AF647), 200 µg/ml, for WB (RGB), IF, IHC(P) and FCM; and to either Alexa Fluor® 680 (sc-518176 AF680) or Alexa Fluor® 790 (sc-518176 AF790), 200 µg/ml, for Near-Infrared (NIR) WB, IF and FCM.

APPLICATIONS

AGL (B-11) is recommended for detection of AGL of mouse, rat and human origin by Western Blotting (starting dilution 1:100, dilution range 1:100-1:1000), immunoprecipitation [1-2 µg per 100-500 µg of total protein (1 ml of cell lysate)], immunofluorescence (starting dilution 1:50, dilution range 1:50-1:500) and solid phase ELISA (starting dilution 1:30, dilution range 1:30-1:3000).

Suitable for use as control antibody for AGL siRNA (h): sc-88368, AGL siRNA (m): sc-140904, AGL shRNA Plasmid (h): sc-88368-SH, AGL shRNA Plasmid (m): sc-140904-SH, AGL shRNA (h) Lentiviral Particles: sc-88368-V and AGL shRNA (m) Lentiviral Particles: sc-140904-V.

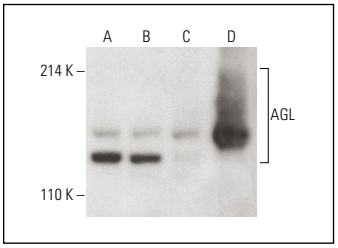
Molecular Weight of AGL: 160 kDa.

Positive Controls: K-562 whole cell lysate: sc-2203, human skeletal muscle extract: sc-363776 or Jurkat whole cell lysate: sc-2204.

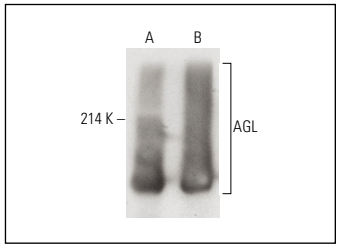
RECOMMENDED SUPPORT REAGENTS

To ensure optimal results, the following support reagents are recommended: 1) Western Blotting: use m-IgGκ BP-HRP: sc-516102 or m-IgGκ BP-HRP (Cruz Marker): sc-516102-CM (dilution range: 1:1000-1:10000), Cruz Marker™ Molecular Weight Standards: sc-2035, UltraCruz® Blocking Reagent: sc-516214 and Western Blotting Luminol Reagent: sc-2048. 2) Immunoprecipitation: use Protein A/G PLUS-Agarose: sc-2003 (0.5 ml agarose/2.0 ml). 3) Immunofluorescence: use m-IgGκ BP-FITC: sc-516140 or m-IgGκ BP-PE: sc-516141 (dilution range: 1:50-1:200) with UltraCruz® Mounting Medium: sc-24941 or UltraCruz® Hard-set Mounting Medium: sc-359850.

DATA



AGL (B-11): sc-518176. Western blot analysis of AGL expression in K-562 (**A**), Jurkat (**B**) and U-698-M (**C**) whole cell lysates and human skeletal muscle tissue extract (**D**). Detection reagent used: m-IgGκ BP-HRP: sc-516102.



AGL (B-11): sc-518176. Western blot analysis of AGL expression in human skeletal muscle (**A**) and mouse skeletal muscle (**B**) tissue extracts. Detection reagent used: m-IgG Fc BP-HRP: sc-525409.

STORAGE

Store at 4° C, **DO NOT FREEZE**. Stable for one year from the date of shipment. Non-hazardous. No MSDS required.

RESEARCH USE

For research use only, not for use in diagnostic procedures.

PROTOCOLS

See our web site at www.scbt.com for detailed protocols and support products.